Congenital heart diseases are among the most common malformations in human fetuses, and, if considered as a group, they are the most frequent. Because of their poor prognosis, they significantly contribute to infant mortality, accounting for approximately 10% of infant deaths and half of the deaths due to congenital malformation.

In 1998, the coefficients of neonatal mortality were 10.0/1000 and 8.3/1000 live births, respectively, for the state of Rio Grande do Sul and the city of Porto Alegre. In 1999, these indices dropped to 9.6/1000 and 7.1/1000 live births for the state of Rio Grande do Sul and the city of Porto Alegre, respectively. These data were provided by the Municipal Secretariat of Health and Social Service and by the State Secretariat of Health of Rio Grande do Sul and have not yet been published.

Neonatal deaths (170/291), particularly those occurring in the first week of life (121/170), accounted for more than half (58.4%) of the infant mortalities in the city of Porto Alegre in 1999. When considering postnatal mortality, 61.2% of the deaths in the period from 28 to 364 days occurred in the first months of life (also unpublished data delivered by the Municipal Secretariat of Health and Social Service and by the State Secretariat of Health of the state of Rio Grande do Sul).

In 1998, the major causes of neonatal mortality in the city of Porto Alegre (194 deaths occurring from 0 to 27 days of life) were, in decreasing order of importance, as follows: respiratory distress syndrome (25 cases), prematurity (24 cases), cardiovascular malformations (23 cases), and fetal repercussions of maternal complications (18 cases), all of which were responsible for approximately 50% of the 194 deaths in that age group. In 1998, perinatal afflictions and congenital anomalies, which were, respectively, the first and second group of most important causes of infant mortality, accounted for 39.5% and 20.7% of infant mortality in the city of Porto Alegre, corresponding to 60.2% of the deaths occurring in infants under the age of 1 year (227 in a total of 377).

Cardiovascular malformations (International Classification of Diseases, 10th review, codes Q20 to Q25) are the major cause of infant death due to congenital malformations, accounting for approximately one third of the deaths in that specific age group, or even for almost one half of the deaths, as in 1999. In that year, 7.9% (23/291) of the total deaths of infants under the age of 1 year in the city of Porto Alegre were due to cardiovascular malformations (unpublished data delivered by the Municipal Secretariat of Health and Social Service and by the State Secretariat of Health of the state of Rio Grande do Sul).

Objective
To study the morphological and functional abnormalities of the fetal cardiovascular system detectable through ultrasonography.

Methods
The study comprised 3,980 fetuses of pregnant women with no obstetrical or cardiological risk in the municipality of Porto Alegre, who, from July 1996 to November 2000, underwent echocardiography for screening fetal cardiovascular malformations. The examinations consisted of 4-chamber and right and left ventricular outflow tract echocardiographic views.

Results
One hundred and three diagnoses of fetal cardiovascular abnormalities were established, corresponding to 2.58% (103/3,980) of the population studied, or 25.8/1000, of which 47 represented morphological and functional changes in the fetal heart with an overall prevalence of 11.8/1000 (47/3,980), and 56 showed changes in refringence (golf ball). Three false negative and no false positive results increased the overall prevalence to 12.5/1000 (50/3,980), or 26.6/1000 (106/3,980), when the refringence changes were also included.

Conclusion
The detection of cardiovascular malformations still in the intrauterine period, aiming at perinatal planning, is possible, which confirms the international experience. Implantation of cardiovascular malformation screening into the routine care network of obstetrical ultrasonography is suggested.

Key words
fetal cardiac abnormalities, prenatal echocardiography, low risk, pregnancies
Studies on the overall prevalence of congenital heart diseases carried out in the postnatal period showed a variation from 3.5 to 13.7/1000 live births. They were performed at different times, in different regions, and with nonstandardized methodologies, which makes accurate comparisons between them impossible.

It is worth stressing that, when studying the overall prevalence of congenital heart diseases, the prevalence in stillborns is approximately 10 times greater than in newborns.16–19

Despite the scarcity of information in the literature about the prevalence of congenital heart diseases in aborted fetuses, it is estimated that, if the congenital heart diseases present in these fetuses and stillborn infants were added to those already known, the overall prevalence of congenital heart diseases would be 5 times greater.22

Although it is important to select groups with increased risk for cardiovascular malformations, only 10% of the newborns with congenital heart disease have an identifiable risk factor during pregnancy.23

Several authors have reported prevalence rates of prenatally diagnosed congenital heart diseases in different countries in the past 15 years.24–33 These prevalence rates have ranged from 3.3 to 14.9/1000 live births,25 and this variation may be attributed to differences in technology and study design.

Although congenital heart diseases represent a small percentage of all heart diseases, they account for a significant share of mortality, morbidity, disability, and potential loss of years with quality of life, affecting mainly patients under the age of 1 year. As far as is known, no study aiming at determining the prevalence of cardiovascular malformations in this population in the city of Porto Alegre exists.

Therefore, a cross-sectional study of prevalence was carried out in part of the geographic area of the municipality of Porto Alegre. This study aimed at determining the overall and specific prevalence of the morphological and functional abnormalities of the fetal cardiovascular system detectable with the aid of 4-chamber, left and right ventricular outflow tract, and aortic arch echocardiographic views in a low-risk pregnant population.

**Methods**

All pregnant women on prenatal follow-up in primary health care units, where low-risk prenatal follow-up is carried out in the municipality of Porto Alegre, were eligible for the study. They were consecutively and randomly recruited between July 1996 and November 2000, and fetuses from pregnant women from other municipalities were not accepted.

Screening of fetal cardiac abnormalities was performed through visualization of the following echocardiographic views: 4-chamber, the right and left ventricular outflow tract, and aortic arch.23,34,35 The following obstetrical echographs were used: 1) Shi medzu 350 with a 3.5-MHz convex transducer with a capacity for 2-dimensional image and M-mode; 2) Aloka 500 with a 3.5-MHz convex transducer with a capacity for 2-dimensional image and M-mode; and 3) Medson Eureka with a 3.5-MHz convex transducer with a capacity for 2-dimensional image and M-mode. In addition, the following equipment of the Fetal Cardiology Unit of the Instituto de Cardiologia/Fundação Universitária de Cardiologia was used: ATL Ultramark-9 Digital Plus, Acuson XP-10, and Acuson Aspens. No routine measurement of the structures identified was performed.

Due to the impossibility of transporting all the pregnant women studied, echocardiography was performed at the primary health care units with an ultrasound device allowing visualization of the fetal heart.

Fetal echocardiograms were provided free of charge as a routine procedure during prenatal care after the 18th gestational week to the pregnant women who spontaneously participated in the study after receiving explanations (approved by the UP 1673/95 protocol of the Committee on Ethics and Research of the Institute). The team of examiners who visited the primary health care units comprised ideally one physician trained in image obtainment (not necessarily an echographer), one secretary, one or more medical students (with scientific initiation grants) or trainees of the Fetal Cardiology Unit, one psychologist, one social worker or trainees in these sectors, who aided in data gathering and in the execution of the projects that were part of the major project of prenatal detection of congenital heart diseases.

The pregnant women who met the criteria for participating in the study and who spontaneously knew about the detection program or those who, due to other reasons, missed the opportunity to undergo the examination in the referral primary health care unit, or those whose gestational ages were more advanced and whose appointment at the referral primary health care unit was scheduled for a date after the probable date of delivery could schedule an appointment at the screening team at the Instituto de Cardiologia facilities.

Concomitantly with fetal echocardiography performance, data on the ongoing pregnancy were obtained (identification of the patient, number of pregnancies and parity, prior medical and obstetrical history, evolution and problems of the current pregnancy) in the search for factors that could signal the risk for fetal cardiovascular malformations (diabetes mellitus; collagenosis; phenylketonuria; use of medications, such as indomethacin, lithium, and phenytoin; smoking; alcohol consumption; family history of or previous pregnancies with congenital heart diseases; congenital infections; arrhythmias; noncardiac malformations; fetal growth delay; hydronephrosis; oligohydramnios; and polyhydramnios).

Once a fetal cardiovascular malformation was suspected, the pregnant woman was immediately referred for echocardiography to the Fetal Cardiology Unit, a tertiary health care unit, with preferential scheduling and a time interval shorter than 1 week. If the existence of a fetal cardiovascular malformation was confirmed, the fetus was included in the protocol of the unit, usually with obstetrical care scheduled at Instituto de Cardiologia facilities.

The follow-up of postnatal outcomes was performed through information obtained from the mothers, from the maternity services, and the services of Neonatology and Pediatric Cardiology, where the babies were born, and from the fetal and infant morbidity and mortality files of the Municipal Secretariat of Health and Social Service of the municipality.

Data collected were stored in a specific database using EPIINFO 6.04b software (Center for Disease Control, Atlanta, GA, USA, and World Health Organization, Geneva, Switzerland, October 1997). The descriptive statistical analysis (frequency, means, medians, and percentages) was conducted from data stored in that database.
Results

From July 1996 to November 2000, 3,980 screening fetal echocardiograms were performed in a population at low obstetric and cardiological risk for cardiac malformations, 3,263 of which were performed in primary health care units and 717 in the facilities of the Fetal Cardiology Unit of the Instituto de Cardiologia (tab. I).

The patients’ ages ranged from 13 to 48 years (mean, 25.2; standard deviation, 6.7; median, 24). The number of pregnancies varied from 1 to 13 (mean, 2.5; standard deviation, 1.7; median, 2). Parity ranged from 0 to 9 (mean, 1; standard deviation, 1.4). The mean maternal educational level was 8 years (the fundamental level). The formal type of wedding predominated (39%), and the median annual income was 3 minimum wages (minimum wage = R$ 151.00).

The gestational age, according to the patient, at which fetal echocardiography was performed ranged from 14 to 42 weeks (mean of 28.6 and standard deviation of 5.7). As 756 pregnant women did not know for sure their gestational ages, the fetal biparietal diameter was measured during echocardiography for estimation of the gestational age which ranged from 8 to 41 weeks (mean of 28 and standard deviation of 6).

In that population, 6.5% of the women reported some previous systemic disease (systemic lupus erythematosus, diabetes mellitus, systemic arterial hypertension, epilepsy, or congenital heart disease), 27.2% reported the use of medications or drugs, or both, during gestation (tobacco, alcohol, lithium, indomethacin, phenytion, acetylsalicylic acid, or oral contraceptives), and 3.6% reported systemic disease during the on-going pregnancy (diabetes mellitus, systemic arterial hypertension, preeclampsia, or eclampsia). Of 51 women reported systemic disease during the on-going pregnancy, 3.6% reported systemic disease (systemic lupus erythematosus, diabetes mellitus, systemic arterial hypertension, or both) during gestation (tobacco, alcohol, lithium, indomethacin, phenytion, acetylsalicylic acid, or oral contraceptives), and 3.6% reported systemic disease during the on-going pregnancy (diabetes mellitus, systemic arterial hypertension, preeclampsia, or eclampsia). Of 51 women reported systemic disease during the on-going pregnancy, 3.6% reported systemic disease (systemic lupus erythematosus, diabetes mellitus, systemic arterial hypertension, or both) during gestation (tobacco, alcohol, lithium, indomethacin, phenytion, acetylsalicylic acid, or oral contraceptives), and 3.6% reported systemic disease during the on-going pregnancy (diabetes mellitus, systemic arterial hypertension, preeclampsia, or eclampsia).

Table I shows that 3,263 pregnant women (82%) underwent a screening fetal echocardiography in primary health care units, and 717 (18%) sought the Fetal Cardiology Unit for the screening test.

Of the 3,263 patients who sought the primary health care units, 2,924 were informed about the need or not to repeat fetal echocardiography. In 2,788 of 3,263 (85.4%) pregnant women, the echocardiogram was conclusive and normal in the first attempt. Most of the time, this was due to early pregnancies or to the position of the conceptus at the time of the first examination. In the same population, the heart could be visualized on the 4-chamber view in 96.7% of the examinations performed. The interventricular septum was identified 94.9% of the time, the left ventricular outflow tract in 94.2% of the fetuses, and the right ventricular outflow tract in 90.4% of the examinations (tab. III). The aortic arch image was obtained 71.6% of the time.

The 4-chamber and the ventricular outflow tract views were visualized in all the 717 fetuses examined in the Fetal Cardiology Unit. In addition, in some instances, the diagnoses were aided by the use of Doppler.

The examiners considered that 17% of the fetuses of the pregnant population originating in the primary health care units had reasons that made the obtainment of the echocardiographic images difficult. The most frequent of these reasons were as follows: position of the conceptus, early gestational age, and maternal obesity. In only 3.3% of the time, the 4-chamber view was not obtained, which hindered mostly the obtainment of the aortic arch image.

Three hundred and thirty-two of 3,263 fetuses were referred for echocardiography in the facilities of the Fetal Cardiology Unit. In that population originating in the primary health care units, the tertiary health care facility, due to abnormalities suspected of or evidenced in the screening fetal echocardiograms performed in the primary health care units, which corresponded to 0.2% of the population studied. Of these 332, 176 (53%) pregnant women scheduled and underwent the examination.

A total of 103 diagnoses of fetal cardiac abnormalities were established (tab. IV), corresponding to an overall prevalence of 25.8/1000 (103/3,980) (95% confidence interval of 21.1 to 31.3). Of these, 81 (81/3,263 or 2.4%) were found among the population originating from the primary health care units and 22 (22/717 or 3.0%) among the pregnant women who sought the Fetal Cardiology Unit for the screening examination. The percentages are not statistically different among themselves (z = 0.80; P = 0.37).

Fifty-six fetuses showed changes in refringence (golf balls), 44 originating from the primary health care units and 12 from the Fetal Cardiology Unit.

<p>| Table I - Population studied (n = 3,980) |</p>
<table>
<thead>
<tr>
<th>Amplitude</th>
<th>Mean (± SD)</th>
<th>Median</th>
</tr>
</thead>
<tbody>
<tr>
<td>Maternal age (years)</td>
<td>13-48</td>
<td>25.2 ± 6.7</td>
</tr>
<tr>
<td>Parities</td>
<td>0-9</td>
<td>1 ± 1.4</td>
</tr>
<tr>
<td>Gestational age - LMD (weeks)</td>
<td>14-42</td>
<td>28.6 ± 5.7</td>
</tr>
<tr>
<td>Gestational age - ECHO (weeks)</td>
<td>8-41</td>
<td>28 ± 6.0</td>
</tr>
<tr>
<td>Educational level (years)</td>
<td>0-15</td>
<td>8.1 ± 2.9</td>
</tr>
<tr>
<td>Familial income (MW)</td>
<td>0-99</td>
<td>4.9 ± 5.8</td>
</tr>
</tbody>
</table>

SD = standard deviation; MW (minimum wage) = R$ 151.00; LMD = last menstrual date; ECHO = echograph

<p>| Table II - Results of the screening echocardiographies according to origin of the pregnant women |</p>
<table>
<thead>
<tr>
<th>PU</th>
<th>FCU</th>
<th>TOTAL</th>
</tr>
</thead>
<tbody>
<tr>
<td>Normal</td>
<td>3,182 (80.9%)</td>
<td>695 (17.5%)</td>
</tr>
<tr>
<td>CVM</td>
<td>37 (0.9%)</td>
<td>10 (0.2%)</td>
</tr>
<tr>
<td>Golf Ball</td>
<td>44 (1.1%)</td>
<td>12 (0.3%)</td>
</tr>
<tr>
<td>Total</td>
<td>3,263 (82.2%)</td>
<td>717 (18.1%)</td>
</tr>
</tbody>
</table>

PU = primary health care unit; FCU = Fetal Cardiology Unit; CVM = cardiac vascular malformations.

| Table III - Percentages of visualization of the fetal echocardiographic views in the pregnant women originating from the primary health care units |
|---|---|---|---|---|
| 4C | IVS | LV | RV | AA |
| Normal | 95.3 | 93.5 | 93.9 | 90.0 | 71.2 |
| Altered | 1.4 | 1.4 | 0.3 | 0.4 | 0.4 |
| Non visualized | 3.3 | 5.1 | 5.8 | 9.6 | 28.4 |

4C = 4-chamber; IVS = interventricular septum; LV = left ventricular outflow tract; RV = right ventricular outflow tract; AA = aortic arch.
Table IV - Diagnoses established: absolute (n) and relative (%) frequency according to the origin of the pregnant women and specific prevalence (per 1000 live births).

<table>
<thead>
<tr>
<th>Diagnoses</th>
<th>PU</th>
<th>FCU</th>
<th>Total</th>
<th>%</th>
<th>Specific prevalence</th>
</tr>
</thead>
<tbody>
<tr>
<td>Arrhythmias</td>
<td>7</td>
<td>3</td>
<td>10</td>
<td>9.7</td>
<td>2.5</td>
</tr>
<tr>
<td>Conotruncal defects</td>
<td>1</td>
<td>1</td>
<td>1</td>
<td>0.9</td>
<td>0.2</td>
</tr>
<tr>
<td>Interventricular septal defect</td>
<td>2</td>
<td>2</td>
<td>4</td>
<td>1.9</td>
<td>0.5</td>
</tr>
<tr>
<td>Interventricular septal defect</td>
<td>10</td>
<td>14</td>
<td>24</td>
<td>13.6</td>
<td>3.5</td>
</tr>
<tr>
<td>Complete atrioventricular septal defect</td>
<td>-</td>
<td>1</td>
<td>1</td>
<td>0.9</td>
<td>0.2</td>
</tr>
<tr>
<td>Pericardial effusion</td>
<td>3</td>
<td>3</td>
<td>6</td>
<td>2.9</td>
<td>0.7</td>
</tr>
<tr>
<td>Tricuspid dysplasia</td>
<td>11</td>
<td>1</td>
<td>12</td>
<td>1.1</td>
<td>0.9</td>
</tr>
<tr>
<td>DORVFT + subaortic I VSD + PS</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>0.9</td>
<td>0.2</td>
</tr>
<tr>
<td>Golf ball</td>
<td>44</td>
<td>12</td>
<td>56</td>
<td>54.3</td>
<td>14</td>
</tr>
<tr>
<td>Left ventricular hypoplasia</td>
<td>2</td>
<td>1</td>
<td>3</td>
<td>2.9</td>
<td>0.7</td>
</tr>
<tr>
<td>Hypertrophic cardiomyopathy</td>
<td>6</td>
<td>6</td>
<td>12</td>
<td>5.8</td>
<td>1.5</td>
</tr>
<tr>
<td>Truncus arteriosus</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>0.9</td>
<td>0.2</td>
</tr>
<tr>
<td>Ventricular tachycardia</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>0.9</td>
<td>0.2</td>
</tr>
<tr>
<td>Left superior vena cava</td>
<td>1</td>
<td>1</td>
<td>2</td>
<td>0.9</td>
<td>0.2</td>
</tr>
<tr>
<td>Total</td>
<td>81</td>
<td>22</td>
<td>103</td>
<td>100</td>
<td>25.8</td>
</tr>
</tbody>
</table>

PU- primary healthcare units; FCU- Fetal Cardiology Unit; DORVFT- double right ventricular outflow tract; I VSD- interventricular septal defect; PS- pulmonary stenosis.

Of the 3,980 echocardiographies, fetal morphological and functional changes were evidenced in 47, representing a prevalence of cardiovascular malformations of 11.8/1000 (95% confidence interval of 8.6 to 15.6).

In both subgroups, the most frequent abnormalities, except for the changes in refringence, were interventricular septal defects, followed by alterations in cardiac rhythm (tab. IV).

No false-positive result was found in the 3,980 fetuses examined, and only 1 died in utero. Of the 87 fetuses, 86 were born alive, but 5 died after birth.

In the search for false-negative results, the only data available were those referring to the years 1998, 1999, and part of 2000 (partial data), representing 2,767 pregnant women.

In the Recording System of Live Births in Porto Alegre, 828 names of mothers of newborns were found in the years 1998 and 2000. The other names could not be found in the recording system of mortality in the municipality, except for 10 with death certificates, whose basic cause of death excluded cardiovascular malformation.

Three false-negative results (one interventricular septal defect and coarctation of the aorta, one pulmonary and aortic stenosis, and one Ebstein’s anomaly) were found, elevating the overall prevalence of fetal cardiovascular malformation to 12.5/1000 (50/3,980; 95% confidence interval of 9.3 to 16.5), or 26.6/1000 (partial data), representing 2,767 pregnant women.

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Discussion

The results obtained during the screening for fetal cardiovascular malformations in the population studied confirmed the international experience of 24-33, with a prevalence of cardiovascular malformations in the prenatal period of 12.5/1000, among which 47 morphological and functional alterations of the fetal heart and 56 alterations in endocardial refringence were identified on screening echocardiography.

Although not sufficiently studied and considered unspecific alterations, golf balls may constitute cardiac markers of systemic or chromosomal diseases, which are frequent findings in 4-chamber view. They occur 3 to 8% of the time, more commonly in the left ventricular papillary muscles, but are also visible in the interventricular septum and in the right ventricle. Histologically, they are an excessive build-up of calcium in the myocardium32.

Of the morphological and functional alterations, interventricular septal defect, diagnosed in 14 fetuses, was the most frequently found disease in our study, with a prevalence of 3.5/1000. These results are consistent with the international literature.

Helfner et al.31, in a study whose design is similar to ours, reported defects in the interventricular septum as the most commonly identified morphological and functional alterations, although only 10 out of 36 had been prenatally diagnosed. Those authors also reported that the prevalence found (13.3/1000) was higher than expected (around 8/1000) and might have been due to a more insistent search for cardiovascular malformations than that carried out by other authors.

Arrhythmias are another group of very common disorders in the intrauterine period, and they may be a potential cause of hydrops, sudden death, and fetal death when not recognized and properly followed up.

The 4-chamber view is the most important for screening cardiovascular malformations, and it was altered in 60% of the greater malformations35.

Although most of the abnormalities affecting the right or left side of the heart cause an abnormal image in the 4-chamber view, the most common defects of the great vessels, such as tetralogy of Fallot and transposition of the great vessels, are frequently associated with a normal image in the 4-chamber view in fetal echocardiography.

Several authors have proposed the addition of the images of the left and right ventricular outflow tract to the image obtained with the 4-chamber view to increase the indices of prenatal detection of cardiovascular defects24,26,27,36,39.

DeVore reported that visualization of the ventricular outflow tracts, although requiring some additional training, is possible in 97% of the fetal echocardiograms. In this study, the right ventricular outflow tract was visualized in 90% of the cases, and the left ventricular outflow tract was visualized in 93.9% of the cases36.

The systematic visualization of the right and left ventricular outflow tracts allowed the additional visualization of 4 (8.9%) heart disease (2 coarctations, 1 interruption of the aortic arch, and 1 double right ventricular outflow tract), and 1 truncus arteriosus, which, if isolated, are theoretically undetectable in the 4-chamber view.

Obtaining of other views, such as the ductal and aortic arches, does not contribute to improvement in the prenatal diagnosis40.

Bronkey et al.37, studying the accuracy of the 4-chamber and right and left ventricular outflow tract views for detecting heart diseases still in the prenatal period, reported that, in the hands of experienced professionals, these views are obtained in more than 95% of the fetuses aged 18 weeks or more, and 83% of the cardiovascular malformations may be identified.

Several studies reported that the 4-chamber view can detect 5 to 81% of the fetal heart defects32,34-45. The addition of the
visualization of the right and left ventricular outflow tracts on echocardiography may increase the index to 14 to 83%.

In our population, the 4-chamber view could not be visualized only 3.3% of the time, the left outflow tract 5.8% and the right ventricular outflow tract 9.6% mainly due to the position of the fetus, early gestational age, or maternal overweight.

The inclusion of Doppler color flow mapping is not essential for the anatomical recognition of abnormal hearts, and diastases may be identified with the isolated 2-diagnostic examination. As this technology significantly increases the costs of equipment acquisition, that evidence reduces the costs of prenatal screening, because it allows the use of a not so sophisticated echocardiography.

Cardiac abnormalities were detected in 3 newborns considered normal on prenatal examination (false-negative). One newborn was identified through the methodology of postnatal information search in the pediatric cardiology and maternity services of the city, with minimal interventricular septal defect and coarctation of the aorta. However, 2 screening examinations considered normal by the team that visited the primary health care unit, but were referred to the Fetal Cardiology Unit in the following weeks due to changes in the obstetrical ultrasonography, still in time for diagnosing their diseases (1 had Ebstein's anomaly, and the other had severe stenosis of the pulmonary artery and aorta) prior to birth. In reality, the diagnosis was established during the intrauterine period, but, on behalf of scientific strictness, they should be considered false-negative, because they were screened and considered normal. The most probable explanation seems to be the precociousness of screening echocardiography (examinations performed at the 18th and 20th gestational weeks).

Usually these anomalies, especially the small atrial and ventricular septal defects, are not visualized in screening echocardiography as 47. According to Leslie et al 48, these defects, along with coarctation of the aorta, are responsible for the greater number of false-negative results.

Of the false-negative results, the first corresponds to a disease with recognized difficulty to be visualized in the screening views and with the basic equipment used. Benacerraf et al 49 diagnosed only 4 fetuses with coarctation of the aorta among the 9 with a suspicion of apparently enlarged right ventricle as compared with the left ventricle on the 4-chamber view. Hornberger et al 50 stressed that the intrauterine diagnosis of coarctation of the aorta is still a challenge, and, in multicenter study designed to identify fetal echocardiographic characteristics that aid its observation, reported that the quantitative hypoplasia of the distal arch (transverse arch and isthmus) is the most valuable finding in predicting coarctation.

Several reports 51 in the literature refer to postnatal clinical control, including those aiming at establishing the prevalence of congenital heart diseases in the prenatal period. The initial studies assessing the role played by the 4-chamber view in the screening of fetal cardiac abnormalities in referral centers and with a non very expressive sample size made the postnatal clinical control, performed by the pediatrician in the follow-up of infants, the method for obtaining information about the false-negative results 52, 53, 54, 55.

The existence of other fetuses with cardiovascular malformations that were not distinguished on screening echocardiography should be considered, because the gold standard for postnatal control was not used, which was absolutely impossible considering the current stage of organization of our medical care in pediatric cardiology. However, this fact only reinforces the observation that the prevalence of cardiovascular malformations should be greater in the prenatal period.

Although our study did not aim at determining the sensitivity (depending on the true number of cardiovascular malformations assessed through the method of cold gold standard) and specificity of the method (depending on the false-positive results), they may be calculated from the results found. As 3 false-negative results were found (raising the total number of cardiovascular malformations of the population studied to 50), the sensitivity was established as 94% (47/50), while the specificity was 100% because no false-positive results existed.

The positive predictive value, which is the assessment of the truly positive results among those with a positive test, also depends on the detection of false-positive results. No fetus diagnosed as having cardiovascular malformations was later identified as normal in the postnatal period; therefore, the positive predictive value of the test in the population studied is 100%.

By identifying 3 false-negative results, the negative predictive value, which is the assessment of the truly negative results among those with a screening considered normal, was 99.9% (3,874/3,877).

Our study was weakened by the methodological decision of not performing color Doppler echocardiography in the postnatal period for controlling all fetuses (recognized gold standard), as well as by not systematically performing an autopsy in all dead fetuses and neonates. However, it has been agreed that the prevalence found here is very close to the real index in the municipality of Porto Alegre.

Even when the results and efficacy of prenatal screening of cardiac abnormalities are questioned, it should be considered that perinatal planning undoubtedly increases the chances of survival of malformed fetuses.

Although almost all cardiac structural defects detectable on the 4-chamber view may result in death in the first years of childhood, infant mortality may be reduced with a prenatal diagnosis 52.

The infant mortality rate in the state of Rio Grande do Sul and, more specifically, in the city of Porto Alegre, where cardiovascular malformations account for 48% of the deaths due to congenital anomalies and for 7.9% of all deaths before the age of 1 year, is very similar to that reported in a study by Abu-Harb et al 57 carried out in the northern region of England. In that study, of 230,654 births, 43% of all deaths were due to congenital malformations and 9% of all infant mortality was due to cardiac malformations. Those authors stressed the fact that only 70% of the heart diseases had been diagnosed prior to death.

In our study, of the 47 morphological and functional alterations found, at least 10 (21.2%) required planning of the management to be adopted in the immediate neonatal period, because of the potential hemodynamic repercussions that they could trigger.

It is worth noting that the system of perinatal medical care, especially in the neonatal one, in the city of Porto Alegre is overloaded with patients from the city, the metropolitan region, and the cities from the inner area of the state, who maintain the occupancy rates of the neonatal intensive care units always saturated.
Newborns without a prenatal diagnosis largely contribute to the chaos in the sector, because most of the services do not have adequate professionals and equipment for establishing the differential diagnosis with congenital heart diseases inside the ICU. The lack of intra- and interhospital transportation, in addition to the time, often vital, which has to be spent for diagnostic elucidation, are factors that add costs and risks to the management of these patients.

Oliveira et al. have already reported that 74% (59/80) of the mothers of newborns with congenital heart diseases admitted to the pediatric intensive care unit of the Instituto de Cardiologia had already undergone at least 1 echographic examination during pregnancy. However, no congenital heart disease was suspected in 54 fetuses (91.6%) on obstetrical ultrasonography, suggesting that the fetal heart was not being systematically assessed by the echographers.

DeVore estimating the costs for detecting 1 heart disease at services with no specific training for prenatal diagnosis in the United States, reported that the creation of a network under the supervision of specialists to increase the number of prenatal diagnoses of cardiovascular malformations may be very productive.

Culturally, we do not easily accept autopsy, and few are the cases diagnosed or confirmed with it. The feasible alternative for determining the prevalence of cardiovascular malformations for us is echocardiography, and the time to perform it is during pregnancy.

Aerts et al. classifying infant deaths in the city of Porto Alegre from 1997 to 1999 according to the basic cause and their potential for reducebility, stressed the amazing percentage of 94.1% (274/291) of deaths before the age of 1 year in 1999 considered avoidable and the inexpresiveness of the immune-preventable deaths in infant mortality (reflecting the great investment in the area so far). Another point is the significant participation of the prenatal component in the potential for reducibility (responsible for 49.4%, or 59/80 newborns, or 84/164 babies), which still needs to be done to obtain a greater control of pregnancies, aiming to reduce infant mortality.

But the highlight in this study is the greatest number of deaths in the group of reducebile deaths with early diagnosis and treatment [141/372 (37.9%) in 1997; 174/377 (46.1%) in 1998; and 127/291 (43.6%) in 1999], representing approximately one third of the neonatal deaths and more than half of the postnatal mortality. Congenital heart diseases are the major group of causes of mortality due to congenital anomalies with an expressive representation in infant mortality in absolute and relative figures, being also responsible for significant morbidity. One of the steps in planning the strategy to fight this scenario and reduce mortality is to articulate the sectors already organized and with the ability to perform this task with the political decision of the public sector of investments in human and technological resources.

According to data gathered, justified reasons exist to consider the municipality of Porto Alegre (and maybe the state of Rio Grande do Sul) mature enough to implement a screening program for fetal cardiovascular malformations.

Our concrete proposal comprises a prenatal follow up with at least 2 obstetrical echographies per each pregnant woman as follows: an early examination to reliably determine the gestational age, and another, after 20-24 weeks, to study the morphology of the conceptus and to follow the pregnancy up. This second examination would include the routine visualization of the fetal heart, among other points of interest, using the 4-chamber and the right and left ventricular outflow tract views. Routine prenatal screening for cardiovascular malformations would be, therefore, feasible on a large scale. The echographers participating in the system to be created and already qualified for professional gynecological and obstetrical practice would receive additional specific training for obtaining and interpreting the fetal heart images under the systematic and continuous supervision of the Fetal Cardiology Unit of the Instituto de Cardiologia of the state of Rio Grande do Sul.

The visualization of a suspicious image, or the mere doubt regarding its presence, would require the pregnant woman and the fetus to undergo fetal echocardiography at a secondary health care unit. The cases confirmed would have the continuity of the follow up by the multi-disciplinary team of the Fetal Cardiology Unit assured, using resources available for intrauterine therapy or planning the perinatal care.

Therefore, we think we would be providing the city with an instrument capable of effective population care, generating scientific knowledge and allowing its growth, and multiplying the experience in a modulated manner. This could be spread to the metropolitan region of Porto Alegre and then to the entire state. It is worth noting that the proposal could be progressively implemented according to the resources available in each place, but with the final objective of effective implementation of a screening network for fetal cardiovascular malformations.

In conclusion, the overall prevalence of morphological and functional abnormalities of the fetal cardiovascular system is detectable using the echocardiographic 4-chamber and right and left ventricular outflow tract views in a low-risk pregnant population in the municipality of Porto Alegre is 12.5/1000. The following specific prevalences were observed: 3.5/1000 for intraventricular septal defect; 2.5/1000 for disordered fetal cardiac rhythm; 1.5/1000 for fetal hypotrophic cardiac myopathy; 0.7/1000 for left ventricular hypoplasia and left cardiac effusion; 0.5/1000 for coarctation or interruption of the aortic arch and pericardial septal defect; and 0.2/1000 for complex heart disease in xiphopagus, tricuspid dysplasia, double outlet ventricular outflow tract with subaortic intraventricular septal defect, truncus arteriosus, ventricular tumor, and left superior vena cava.

There is still a long way to go in regard to collective health, but it is undeniable that approximately 50% of the prevalent or avoidable infant deaths in the city of Porto Alegre depend on investments in technologies and human resources capable of facing this challenge.
References


